Case Report
JCAM

Preoperative Pulmonary Valvuloplasty in Tetralogy of Fallot with Right-To-Left Shunt

Sağdan-Sola Şantlı Fallot Tetralojisinde Preoperatif Pulmoner Valvüloplasti

Barış Buğan[1], Turgay Çelik[2], Uygar Çağdaş Yüksel[2], Serdar Fırtına[3], Atila İyisoy[2]
[1]Malatya Military Hospital, Cardiology Service, Malatya, [2]Gulhane Military Medical Academy, Department of Cardiology, Ankara,
[3]Erzincan Military Hospital, Cardiology Service, Erzincan, Türkiye

Özet

Fallot tetralojisi en sık saptanan siyanotik konjenital kalp hastalığıdır ve sağ ventrikül çıkış yolu obstrüksiyonu, ventriküler septal defekt, ata binen aorta ve sağ ventrikül hipertrofisi ile karakterizedir. Sağ ventrikül çıkış yolu obstrüksiyonu ve ventriküler septal defekt, sendromun majör klinik parçalarıdır. Çoğu hastaya düzeltici bir operasyon uygulanmasına rağmen, Fallot tetralojili hastaların önemli olan az bir kısmına preoperatif palyatif bir işlem uygulanır. Burada biz sağdan-sola şant nedeniyle, düzeltici cerrahi için ameliyat edilemez olarak düşünülen Fallot tetralojili 19 yaşında bir bayan hastada, pulmoner darlığın başarılı palyatif perkütanöz balon valvüloplastisini sunuyoruz.

Anahtar Kelimeler

Balon Valvüloplasti; Sağ-Sol; Şant; Fallot Tetralojisi

Abstract

Tetralogy of Fallot is the most common cyanotic congenital heart disease and characterized by right ventricular outflow tract obstruction, ventricular septal defect, overriding aorta, and right ventricular hypertrophy. Right ventricular outflow tract obstruction and ventricular septal defect are the major clinical components of the syndrome. Although most have undergone a corrective operation, an important minority of patients with tetralogy of Fallot have had a preoperative palliative procedure. Herein we reported a succesful palliative percutaneous balloon valvuloplasty of pulmonary stenosis at an 19-year-old female patient with tetralogy of Fallot who was considered as inoperable for corrective surgery due to right -to-left shunt.

Keywords

Balloon Valvuloplasty; Right-to-Left; Shunt; Tetralogy of Fallot

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Corresponding Author: Barış Buğan, Malatya Military Hospital, Cardiology Service, Malatya, Turkey.
T.: +905323476096 F.: +90 4223362043 E-Mail: bbugan@hotmail.com

Introduction

There is a wide clinical aspect of disease in tetralogy of Fallot (TOF), depending on the severity and localization of the right ventricular outflow truct obstruction (RVOTO) [1, 2]. The ventricular septal defect (VSD) is generally large and non-restrictive. The direction and magnitude of flow through the defect depends on the severity of the RVOTO or increased pulmonary vascular resistance causing Eisenmenger Syndrome. If obstruction to the right ventricular outflow is severe, or if there is an increased pulmonary vascular resistance, a large right-to-left shunt with low pulmonary blood flow and severe cyanosis are present [3].

Recently most patients could undergo complete primary repair during infancy, however balloon dilatation of RVOTO in symptomatic patients with diminutive pulmonary arteries has been reported as a palliative step to prevent repeated surgeries for TOF before corrective surgery [2, 4]. There are concerns regarding to the efficacy and safety of this technique, especially in patients with TOF and right-to-left shunt [4]. In this paper, we reported a 19-year-old female patient with TOF and right-to-left shunt who underwent balloon dilatation of pulmonary stenosis before complete corrective surgery.

Case Report

A 19-year-old female patient with TOF was referred to our centre from Afghanistan for further evaluation. Her symptoms were fainting, dizziness, fatigue, and palpitation. On admission, she was severe cyanotic and had clubbing at her fingers. There was a loud 3/6 systolic murmur at mesocardia and pulmonary focus with a splitting of second heart sound. Electrocardiogram (ECG) showed sinus tachycardia with right bundle branch block and right axis deviation. She was hypoxic at blood gases, and her oxygen (O2) saturation was almost 30 % with room air and 73 % with 5 liter O2 per minute. During mobilization, she has had fainting spells. Biochemical tests were found normal except hemoglobin (Hb) and hematocrit (Htc) values (Hb: 18.2 gr/dl, and Htc: 63.7%). Echocardiograpy revealed right ventricular hypertrophy, overriding aorta, VSD (Figure1) and pulmonary valve stenosis with a peak gradient of 100 mmHg. For further evaluation, cardiac catheterization was performed. Overriding aorta was visualized with right to left shunt after opaque injection. Pulmonary angiography showed that major branches of pulmonary artery were enough developed, and distal bed was intact but thin (Figure 2). Pressure measurements were as follows; a systolic gradient of 112 mmHg at pulmonary valve (pulmonary artery; 18,12,15 mmHg; right ventricule; 130,10,50 mmHg). Pulmonary balloon valvuloplasty was performed succesfully (Tyshsca 22mmX 4) and shunt reversed left to right (Figure 3). Her O2 saturation in blood gases raised above %90 with room air. She was referred to surgery for complete repair of TOF two weeks later.

Discussion

The natural history of TOF is variable and depends on the severity of RVOTO. Twenty-five percent of infants with severe obstruction not treated surgically die within the first year. Left untreated, 40% die by age 3 years, 70% by age 10 years, and 95% by age 40 years. The risk of death is greatest in the first year of life. Major causes of death in surgically untreated patients include hypoxic spells (62%), cerebrovascular accidents (17%), and brain abscesses (13%) [1, 3].

There are many conflicts about the optimum time and appropriate procedure for symptomatic patients with TOF. Although complete corrective surgery should be performed immediately after the diagnosis, the best age for repair is 3–6 months of age [3, 5]. Early age and using trans-annular patch to relieve the RVOTO are high risk predictors for postoperative mortality. Balloon dilatation is an effective and recommended palliative therapy for these patients to postpone the surgery [5]. Balloon dilatation in TOF was first performed Lababidi in 1983 [6]. In 1991 Sreeram et al [4] reported 86% of success rate on series of patients with TOF. Balloon dilatation increases pulmonary blood flow and oxygen saturation and should improve growth of pulmonary artery branches. Pulmonary arterial hypoplasia is major obstacle to perform this therapy. If the pulmonary tree

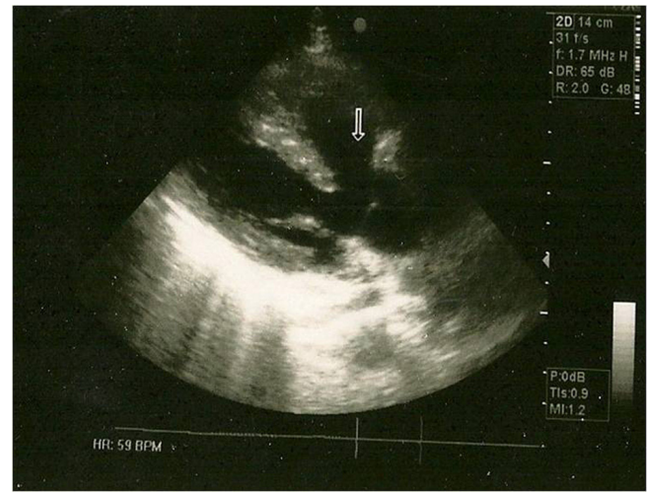

Figure 1. Echocardiogram showing ventricular septal defect and overriding aorta (arrow).

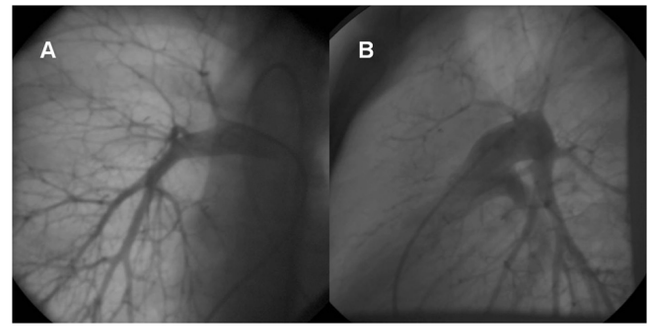

Figure 2. Right(A) and left (B) pulmonary angiography showing pulmonary arterial tree. Distal pulmonary bed is intact but thin.

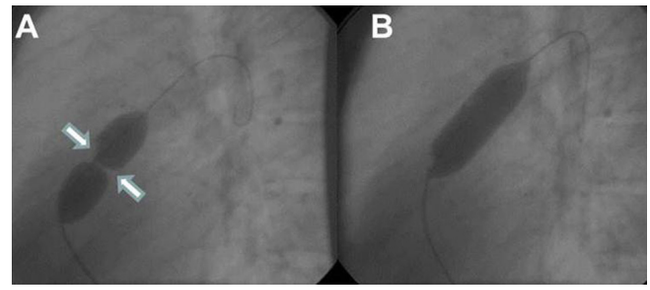

Figure 3. Angiograms showing pulmonary balloon valvuloplasty. Arrows indicating the stenotic pulmonary valve (A). After balloon valvuloplasty stenosis was improved (B).

does not develop enough, acute right heart failure may occur due to volume and pressure overload. So, complete assessment of the pulmonary tree and right ventricular function are very critical [2, 4]. On the other hand, detection of the shunt direction is critical in especially adult patient to detect whether the patient is inoperable or not. If the right to left shunt depends on the severity of the RVOTO, it reverse with ballon dilatation therapy and complete corrective surgery may be performed successfully as our patient.

In conclusion, balloon dilatation is an effective and useful palliative procedure to be a bridge therapy for complete corrective surgery in both early age symptomatic patients and adult patients with right to left shunt, however the physicians should keep in mind that assessment of the pulmonary tree, RV function and the shunt direction are very critical for success of the procedure.

Competing interests

The authors declare that they have no competing interests.

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